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CEREBRAL DISEASE.

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Muscular atrophy in diseases of the nervous system is almost invariably due to lesions affecting the cells of the anterior cornua or the nerves leading from them. Clinically such cases are known as atrophic paralysis, and form a distinct and well defined group. In cerebral disease, or in lesions of the fibres of the cord above the anterior cornua, atrophy is usually absent, although a moderate degree of wasting has often been noted in old standing cases of hemiplegia with contractions and is regarded as due to disease. Of late years, however, cases have been observed in which muscular atrophy has been early noted in limbs paralyzed from various forms of cerebral disease. Contrary to expectation, in some of these no changes have been found either in the anterior cornua or in the peripheral nerves. With the object of drawing attention to this condition I submit the two following cases :

CASE I. Tumour of optic thalamus—hemiplegia—atrophy of muscles of hand, forearm and leg—anterior cornua and peripheral nerves normal.

Miss F., æt. 55, was first seen June 21st, 1892. For a month or five weeks past states that she has had pain over the anterior part of the scalp, intermittent, not severe and present chiefly in the morning. She has also had pain in the back of the neck, most marked on rising from the recumbent posture. About the same time she began to experience difficulty in walking, having a tendency to fall backwards and her knees giving way under her. She has vomited once or twice half an hour after her evening meal, without effort or nausea. There has been giddiness. Her friends state that she has been growing stout since her illness began, and that her memory has been failing for three months.

Present Condition.—The patient is well nourished and somewhat stout. Intelligence is fair, but she exhibits no anxiety about her condition, and there is a tendency to laugh easily.

In walking she moves the legs slowly and requires assistance. There is a marked tendency to fall backward and to the left side.

She is unable to use the hands even for eating. The arms and legs both show a marked degree of muscular power.

The left labio-nasal fold is not so prominent as the right, and the movements in the lower part of the face are not quite so marked as on the right, but the upper muscles of the face move normally. The tongue is protruded straight.

¹ Read before the Canadian Medical Association, at Montreal, August 28, 1896.

The sense of position is normal and there is no ataxia of arms and legs.

Sensation in the face and limb is normal. Both knee jerks are exaggerated, but there is no ankle clonus.

The left eye is shrunken and functionless, the result of an old injury. The right optic disc shows a marked grade of neuritis. The pulse is 90. The heart, lungs and urine are normal.

Mental failure and weakness progressed rapidly, and on July 12th she was admitted to the Montreal General Hospital. She was then quite unable to give any account of herself. She lies on her back with her head slightly retracted, sleeping a great deal day and night. She answers questions only in monosyllables and in slow measured tones after a pause of some seconds, relapsing into a soporose condition when left alone. There is no pain. Urine and faeces are passed in bed.

The left arm and leg are paralysed and flaccid. She is not able to register with the dynamometer with the left hand, but registers 30 with the right. There is some loss of power in the right limbs.

July 15. Urine alkaline s.g 1030 : no albumen ; no sugar. Complaints of pain in right sterno-mastoid muscle.

July 20. She lies in the same condition, sleeping most of the time, and snoring loudly. The pain in the neck has gone. There is not much change from day to day.

July 26. Wasting of the left thenar eminence was noticed to-day, and on measuring the following results were obtained :

	Right.	Left.
Arm.....	9½ in.	9½ in.
Fore-arm.....	8	7½
Hand over thenar eminence.....	7	6½

There was an inch difference in the legs in favour of the right side. No tenderness of the muscles or nerve trunks present. With the faradic battery a much stronger current was required to produce contraction of the muscles of the left hand and forearm, and in the respiratory muscles of the face. In the leg there was slight diminution of electrical irritability to faradism. The galvanic reactions were not obtained. The atrophy progressed rapidly in the hand, and the thenar and hypothenar eminences became flat and depressed. On Aug. 3rd, she passed into a comatose condition, the breathing became rapid and stertorous, and death occurred on the following day (Aug. 4.)

The measurements of the limbs taken after death were as follows :

	Right.	Left.
Arm.....	9½	9½
Fore-arm.....	8	7½
Hand.....	7	6½
Thigh.....	14½	14½
Calf.....	11	10½

During her stay in hospital the temperature was slightly elevated, 98 to 100, and 102½ for three days preceding her death. The pulse was also increased in rate, varying from 72 to 104.

The clinical diagnosis was tumour of the brain, based on the optic neuritis, with other cerebral symptoms. Its locality was regarded as being in the neighbourhood of the cerebellum, owing to the peculiar gait. In the absence of any signs of peripheral neuritis, such as tenderness or loss of sensation, the atrophy of the hand was regarded as due to changes in the anterior cornua of the cord.

AUTOPSY.—*Brain.*—The floor of the third ventricle was full and bulging. The right optic thalamus was enlarged and infiltrated with a white tumour of the same colour as the white substance of the cerebrum; its boundaries were ill-defined and infiltrating. It was rather firmer than the surrounding brain tissue and extended back as far as the superior vermiform process of the cerebellum. The tumour was

placed in Müller's fluid for examination, but during changes in the laboratory was unfortunately lost before its exact boundaries or microscopic characters were determined.

The spinal cord and a portion of the median nerve at the elbow were also removed and placed in Müller's fluid for examination. Sections of the cord at the levels of the 4th, 5th, 6th, 7th and 8th cervical and 1st dorsal nerve were subsequently made and stained by Weigert's method and with carmine. The cells of the anterior cornua presented no diminution in size or number; there was no descending degeneration and the cord was in all respects normal. The median nerve also showed no sign of degeneration.

The abdominal and thoracic organs were under-weight, but otherwise presented no change of importance.

The chief features of this case were hemiplegia and wasting of the paralysed muscles. The wasting was noted fourteen days after the limbs became paralysed. It affected chiefly the thenar muscles of the hand, where the atrophy was considerable, and to a less extent the forearm and leg. The limbs were flaccid and there had never been any irritative symptoms. The wasting in the hand was such as to suggest a lesion of the anterior cornua. Anatomically, however, no microscopic changes were found in the lower motor segment or even in the pyramidal tracts of the cord. During life there was no evidence of neuritis, sensation having been normal, and no tenderness of the nerve trunks or muscles was present.

CASE II.—Sarcoma of crus cerebri—Hemiplegia and rapid atrophy of muscles of hand, forearm, arm and shoulder—Autopsy.

I am indebted to Dr. James Stewart, of Montreal, for brief clinical notes of the following case.

Mr. R., *et.* 47. The first symptom noted was loss of colour vision. He then suffered from severe pain in the head. Weakness in the left arm and leg, gradually increasing in intensity, set in. The muscles of the thenar and hypothenar eminences, the forearm, the arm, the deltoid and lower portion of the pectoralis major wasted rapidly and death occurred four months from the onset of symptoms.

Autopsy.—The thenar and hypothenar eminences, the muscles of the forearm and arm, the deltoid and lower portion of the pectoralis major were much wasted on the left side. On removing the brain a greyish, soft, flattened growth lying on and adherent to the right crus cerebri was observed. The growth reached from the anterior border of pons forward to about the level of a line through the middle of the temporo-sphenoidal lobe. The growth was quadrilateral in shape, $1\frac{1}{2}$ inches long and $1\frac{1}{2}$ inches broad. The third and fourth nerves lay alongside the tumour, whilst the optic tract lay beneath the growth. None of the cranial nerves were involved, a fact which caused much obscurity in localising the growth during life.

Microscopically the tumour proved to be a sarcoma with large vascular spaces. The upper part of the spinal cord was removed and also a portion of the ulnar nerve. Sections of the cord at various levels in the cervical region down to and including the first dorsal segment showed the cells of the anterior cornua to be perfectly normal. There was no degeneration of the lateral columns. The sections were stained both with carmine and by Weigert's method.

Sections of the ulnar nerve were normal. The muscle was not examined.

The chief interest in this case lies in the fact that a considerable degree of atrophy of the muscles of the arm was present, associated with a tumour of the crus cerebri and without lesions of the anterior cornua or peripheral nerves to account for it.

Atrophy of the muscles is occasionally seen in old cases of hemiplegia with contracture, and lesions in the anterior cornua or in the peripheral nerves have been demonstrated. Charcot first described atrophy in the cells of the anterior cornua at levels corresponding with the wasted muscles. Déjerine found degeneration of the peripheral nerves and regards this as the sole cause of the atrophy.

There is, however, a class of cases in which wasting occurs early in the paralysed members and in which no changes either in the anterior cornua or peripheral nerves have been present. The wasting cannot be attributed to disuse, as it occurs too early; and again, it may be present to a considerable extent in muscles only partially paralysed.

Anatomical Lesions.—The pathological conditions in the brain vary both in site and character. In a considerable proportion of the cases tumours have been present, but in others softening or hæmorrhage have existed. Bremer and Carson¹ have collected six (including their own) cases in which a tumour was present. Quincke² has reported a seventh and quotes a case of Barresi's and one of Gliky's. Packard, in a paper read at the meeting of the Pædiatric Society in Montreal, 1896, reported a case of a tumour in a child associated with considerable atrophy, and in both my own cases a cerebral growth was present.

In Babinski's³ case a focus of softening in the centrum ovale minus, in the course of the psycho-motor fibres was found. Eisenlohr, reports two cases, in one of which a recent, and in the other an old, hæmorrhagic focus in the brain was found.

The site of the lesions varies, but all involve some portion of the motor tract. A considerable number of the cases of tumour have been in the motor cortex, but in others the paralysis and ensuing atrophy have resulted from disease of the motor tract in the sub-cortical region and in the internal capsule. The optic thalamus has also been primarily involved with damage to the adjacent internal capsule.

Secondary degeneration of the pyramidal tracts and medulla sometimes occurs, and also degeneration of the opposite cross pyramidal tract of the cord and of the direct pyramidal tract on the same side as the lesion. The degeneration of the pyramidal tracts is by no means constant and it can therefore have nothing to do with atrophy of the muscles.

The most surprising and important fact in these cases of early muscular atrophy is, however, the absence of changes in the motor cells of the anterior cornua, in the anterior nerve roots and in the peripheral nerves. This fact is all the more remarkable inasmuch as

the degree of atrophy is often considerable and occurs within a very short period of time.

The following writers (i.e.) report cases of early muscular atrophy with integrity of the lower motor segment, determined by microscopic examination of the cord and nerves. Quinke, Babinski, Eisenlohr, (two cases), Bremer and Carson which, with my own cases, make a total of seven.

The muscles in the few cases in which they have been examined present changes similar to those found following affections of the nerves (v. Babinski, Eisenlohr i.e.).

No very satisfactory explanation of muscular atrophy with integrity of the lower motor segment has yet been offered. Quinke suggests the presence of trophic centres in the cortex, but were this the case early atrophy might be expected to occur much more frequently. Babinski (i.e.) and Joffrey and Achard⁵ suggests that the motor cells of the anterior cornua undergo dynamic changes, sufficient to interfere with the nutrition of the muscles, but not evidenced by anatomical changes.

Symptoms—The period elapsing between paralysis of the muscles and atrophy varies considerably. It is often difficult to fix owing to the fact that wasting is present when the patient first comes under observation and has not previously been noticed by him, and again, its onset may not be observed by the physician until it has reached a considerable degree. The most rapid onset is recorded by Borgherini⁶ in which muscular atrophy (amounting to a difference of 1 cm. in the arm, 5 cm. in the forearm, 5 cm. in the thigh and 1 cm. in the leg) was noted on the third day after an attack of hemiplegia, but it is usually observed about three⁷ or four⁸ weeks after the onset of paralysis. Although paralysis is usually complete, atrophy may occur where paresis⁹ only is present.

The muscles of the arm usually present an earlier and greater degree of wasting than the leg. The muscles of the arm, forearm, shoulder and hand, may all be affected, and in one of Quinke's cases wasting began in the shoulder and arms, the thigh and calf. When the hand is affected, the atrophy may reach a considerable degree, the eminences of the thenar and hypothenar groups being flat or depressed, and the first interosseous muscle also showing a considerable degree of atrophy.

The difference in the size of the limbs varies from 4 cm. to 5 cm. In the hands, although the difference in measurement may be slight, the atrophy seems to reach at times a higher grade than in other muscles. From these facts it appears that the muscles most affected

are those which suffer most in cerebral paralysis, and in which the movements performed are complex and highly differentiated.

The knee-jerks in the cases under discussion are usually increased and foot clonus has been noted, facts which support the view of integrity of the lower motor segment.

Electrical Reactions.—In the few cases examined the electrical reactions have shown diminished faradic contractility corresponding to the wasting (Quinke). In my first case, however, the faradic irritability was much lowered and strong currents were required to produce contraction. The galvanic irritability has shown slight qualitative and quantitative changes, but never the slow muscular movements seen in nerve degeneration.

Summarising we may state that in a certain small proportion of cases of cerebral disease muscular atrophy occurs early and if present in the hand may reach a considerable degree, and that in these cases no anatomical changes are observable in the lower motor segment.

¹ Bremer and Carson, *Am. Jour. Med. Sc.*, 1851, 1, 133.

² Quinke, *Deut. Arch. Klin. Med.*, 42, 492.

³ Babinski, *C. r. de la Société de Biologie*, 1886, 76.

⁴ Eisenlohr, abstract in *Virchow's Jahresbericht d. Ges. Med.*, 1894, 2, 130.

⁵ *Arch. de Méd., Ecp.*, 1891.

⁶ *Deut. Arch. Klin. Med.*, 45.

⁷ Eisenlohr l. c.

⁸ Eisenlohr, Quinke l.c. [three cases.]

⁹ Quinke l.c., Case I. Bremer and Carson l.c.